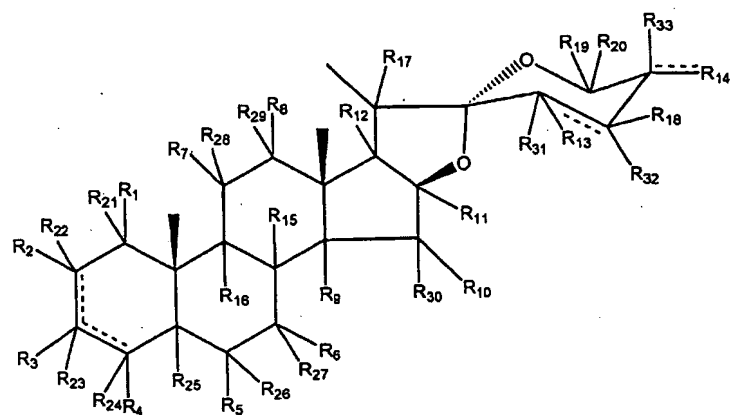


CLAIMS

1. Use of one or more active agent selected from:

5

A. compounds of Formula I:



10

(I)

wherein in the general formula (I):

- R₁, R₂, R₃, R₄, R₅, R₆, R₇, R₈, R₁₀, R₁₃, R₁₈, R₁₉, R₂₀, R₂₁, R₂₂, R₂₃, R₂₄, R₂₆, R₂₇, R₂₈, R₂₉,

15 R₃₀, R₃₁, R₃₂ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;

- R₉, R₁₁, R₁₂, R₁₄, R₁₅, R₁₆, R₁₇, R₂₅, R₃₃ can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;

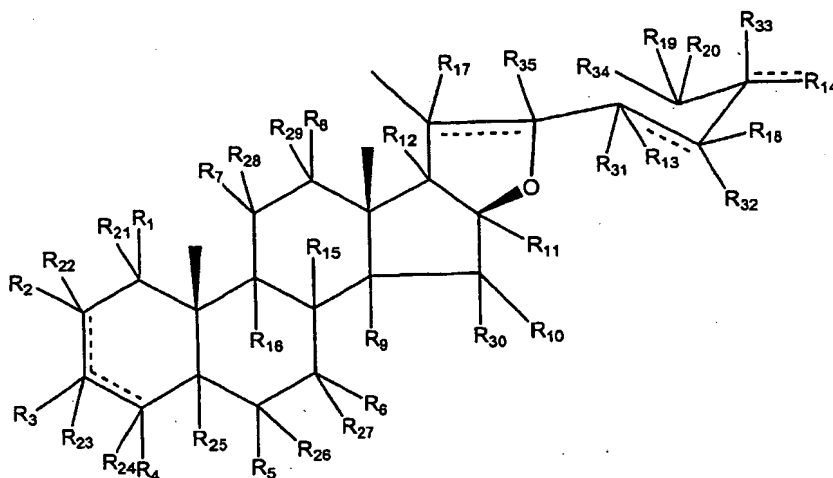
20 represents an optional double bond,

wherein in addition to the above

- either R₃₃ or R₁₄ = alkyl group;

B. compounds of Formula II:

25



(II)

wherein in the general formula (II):

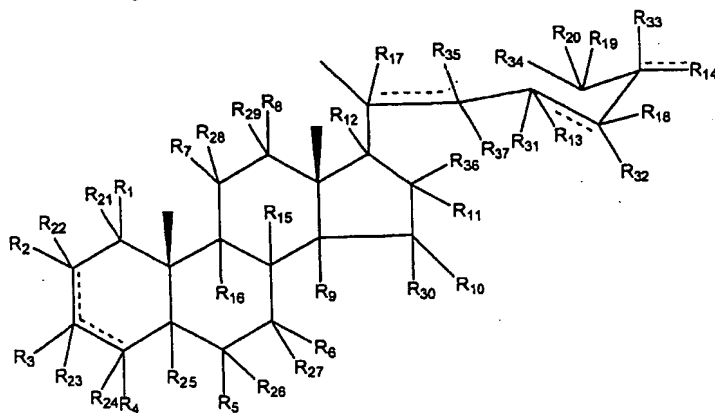
- $R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29},$
- 5 $R_{30}, R_{31}, R_{32}, R_{34}$ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;
- $R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33}, R_{35}$ can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or
- 10 absent;

..... represents an optional double bond.

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group;

- 15 C. compounds of Formula III:



(III)

wherein in the general formula (III):

- R₁, R₂, R₃, R₄, R₅, R₆, R₇, R₈, R₁₀, R₁₃, R₁₄, R₁₈, R₁₉, R₂₀, R₂₁, R₂₂, R₂₃, R₂₄, R₂₆, R₂₇, R₂₈, R₂₉, R₃₀, R₃₁, R₃₂, R₃₃, R₃₄, R₃₅, R₃₆, R₃₇ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;

-R₉, R₁₁, R₁₂, R₁₅, R₁₆, R₁₇, R₂₅ can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R₃₃ or R₁₄ = alkyl group, and

the stereochemistry of R₂₅ is in the β orientation;

D. sapogenin derivatives bearing at least one X radical substituent,

wherein X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO₂-),

- N₃-, NH₂-, MeSO₂NH-, and

- alkyl; and

E. derivative forms of any of the above compounds, in which the carbon atom at the 3-position or, in the case of Formulae II and III, the 3-position carbon atom, the 26-position or each of the carbon atoms at the 3- and 26-positions, carries an O-sugar moiety wherein the sugar group is a mono-, di- or tri-saccharide;

all their stereoisomers and racemic mixtures, all their pharmaceutically acceptable prodrugs and salts, and all mixtures and combinations thereof

in the treatment or prevention of, or in the preparation of compositions for the treatment or prevention of, (i) non-cognitive neurodegeneration, (ii) non-cognitive neuromuscular degeneration, (iii) motor-sensory neurodegeneration, or (iv) receptor dysfunction or loss in the absence of cognitive, neural and neuromuscular impairment, in human and non-human animals suffering therefrom or susceptible thereto.

2. A use according to claim 1, wherein the active agent, or at least one of the active agents, is selected from:

- a. Compounds of the above general formula I, wherein:
- $R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}$ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;
 - $R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33}$ can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;
 - represents an optional double bond,
- 10 wherein in addition to the above
- either R_{33} or R_{14} = alkyl group,
 - and the stereochemistry of R_{25} is in the β orientation;
- * b. Compounds of the above general formula I, wherein:
- 15 - $R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}$ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;
- $R_9, R_{12}, R_{15}, R_{16}, R_{17}$ = H,
- 20 - $R_{11}, R_{14}, R_{25}, R_{33}$ can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl or absent or OR where R = alkyl or acyl group;
- represents an optional double bond
- wherein in addition to the above
- either R_{33} or R_{14} = alkyl group,
- 25 and the stereochemistry of R_{25} is in the β orientation;
- c. Compounds of the above general formula I, wherein:
- $R_1 = R_2 = R_4 = R_5 = R_6 = R_7 = R_8 = R_{10} = R_{11} = R_9 = R_{12} = R_{13} = R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21} = R_{22} = R_{23} = R_{24} = R_{25} = R_{26} = R_{27} = R_{28} = R_{29} = R_{30} = R_{31} = R_{32} = R_{33} = H,$
- 30 - either R_{33} or R_{14} = CH₃
- represents a single bond,
- the methyl group at C25 may be either in the R or S configuration
 - the stereochemistry of R_{25} is in the β orientation and
- wherein in addition to the above
- 35 at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O, and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,
 - (Me-S-), (Me-SO-), (Me-SO₂-), and
 - N₃-, NH₂-, MeSO₂NH-
- 5 - alkyl;

d. Compounds of the above general formula I, wherein:

- R₁=R₂=R₄=R₅=R₆=R₇=R₈=R₁₀=R₁₁=R₉=R₁₂=R₁₃=R₁₅=R₁₆=R₁₇=R₁₈=R₁₉=R₂₀=
 R₂₁=R₂₂=R₂₃=R₂₄=R₂₅=R₂₆=R₂₇=R₂₈=R₂₉=R₃₀=R₃₁=R₃₂=H,
 10 -R₁₄=R₃₃=CH₃,

..... represents a single bond,

- the stereochemistry of R₂₅ is in the β orientation and

wherein in addition to the above

at least one of R₃ or R₂₃ is a X radical, the possible remaining substituent being H, OH, =O,

- 15 and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,
 - (Me-S-), (Me-SO-), (Me-SO₂-), and
 - N₃-, NH₂-, MeSO₂NH-
- 20 - alkyl;

e. Compounds of the above general formula II, wherein

- R₁, R₂, R₃, R₄, R₅, R₆, R₇, R₈, R₁₀, R₁₃, R₁₈, R₁₉, R₂₀, R₂₁, R₂₂, R₂₃, R₂₄, R₂₆, R₂₇, R₂₈, R₂₉,
 R₃₀, R₃₁, R₃₂, R₃₄ are, independently of each other, either H, OH, =O, halo atom, (Me-S-),
 25 (Me-SO-), (Me-SO₂), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or
 absent;

- R₉, R₁₁, R₁₂, R₁₄, R₁₅, R₁₆, R₁₇, R₂₅, R₃₃, R₃₅ can be either a H, OH, halo atom, (Me-S-),
 (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or
 absent;

- 30 represents an optional double bond,

wherein in addition to the above

- either R₃₃ or R₁₄ = alkyl group,

and the stereochemistry of R₂₅ is in the β orientation;

- 35 f. Compounds of the above general formula II or carbohydrate derivatives thereof,
 wherein:

- $R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}$ are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;
- 5 - $R_9, R_{12}, R_{15}, R_{16}, R_{17} = H$,
 - R_{34} = either H, OH, =O, and OR where R = alkyl, acyl or carbohydrate and
 - $R_{11}, R_{14}, R_{25}, R_{33}, R_{35}$ can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;
 represents an optional double bond,
- 10 wherein in addition to the above
 - either R_{33} or R_{14} = alkyl group,
 and the stereochemistry of R_{25} is in the β orientation;
- g. Compounds of the above general formula II or carbohydrate derivatives thereof,
 15 wherein:
 - $R_1 = R_2 = R_4 = R_5 = R_6 = R_7 = R_8 = R_{10} = R_{11} = R_9 = R_{12} = R_{13} = R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21} = R_{22} = R_{23} = R_{24} = R_{25} = R_{26} = R_{27} = R_{28} = R_{29} = R_{30} = R_{31} = R_{32} = R_{33} = H$,
 - $R_{14} = CH_3$,
 - $R_{34} = -OH$ or $-OR$ where R = alkyl, acyl or carbohydrate and
 20 $R_{35} = H$ or is absent
 represents an optional double bond, and
 - the methyl group at C25 may be either in the R or S configuration and
 and the stereochemistry of R_{25} is in the β orientation
 wherein in addition to the above
- 25 at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O,
 and OR where R = alkyl or acyl group or absent,
 and X is chosen from the group consisting of :
 - halo atom,
 - (Me-S-), (Me-SO-), (Me-SO₂-), and
 30 - N₃-, NH₂-, MeSO₂NH-
 - alkyl;
- h. Compounds of the above general formula II or carbohydrate derivatives thereof,
 wherein:
 35 - $R_1 = R_2 = R_4 = R_5 = R_6 = R_7 = R_8 = R_{10} = R_{11} = R_9 = R_{12} = R_{13} = R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21} = R_{22} = R_{23} = R_{24} = R_{25} = R_{26} = R_{27} = R_{28} = R_{29} = R_{30} = R_{31} = R_{32} = H$,

- R₁₄ = R₃₃ = CH₃,
 - R₃₄ = -OH or -OR where R = alkyl, acyl or carbohydrate and
 R₃₅ = H or is absent
 represents an optional double bond, and
- 5 the stereochemistry of R₂₅ is in the β orientation and
 wherein in addition to the above
 at least one of R₃ or R₂₃ is a X radical, the possible remaining substituent being H, OH, =O,
 and OR where R = alkyl or acyl group or absent,
 and X is chosen from the group consisting of :
- 10 - halo atom,
 - (Me-S-), (Me-SO-), (Me-SO₂-), and
 - N₃-, NH₂-, MeSO₂NH-
 - alkyl;
- 15 i. Compounds of the above general formula III, wherein:
 - R₁, R₂, R₃, R₄, R₅, R₆, R₇, R₈, R₁₀, R₁₃, R₁₄, R₁₈, R₁₉, R₂₀, R₂₁, R₂₂, R₂₃, R₂₄, R₂₆, R₂₇, R₂₈,
 R₂₉, R₃₀, R₃₁, R₃₂, R₃₃, R₃₄, R₃₅, R₃₆, R₃₇ are, independently of each other, either H, OH, =O,
 halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R =
 alkyl or acyl group, or absent;
- 20 -R₉, R₁₁, R₁₂, R₁₅, R₁₆, R₁₇, R₂₅ can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-
 SO₂), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;
 represents an optional double bond,
 wherein in addition to the above
 - either R₃₃ or R₁₄ = alkyl group, and
- 25 the stereochemistry of R₂₅ is in the β orientation;
- j. Compounds of the above general formula III or carbohydrate derivatives thereof,
 wherein:
- 30 - R₁, R₂, R₃, R₄, R₅, R₆, R₇, R₈, R₁₀, R₁₃, R₁₄, R₁₈, R₁₉, R₂₀, R₂₁, R₂₂, R₂₃, R₂₄, R₂₆, R₂₇, R₂₈,
 R₂₉, R₃₀, R₃₁, R₃₂, R₃₃, R₃₅, R₃₆, R₃₇ are, independently of each other, either H, OH, =O, halo
 atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or
 acyl group, or absent;
 - R₉, R₁₂, R₁₅, R₁₆, R₁₇ = H,
 - R₃₄ = H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-,
 35 alkyl, OR where R = alkyl, acyl or carbohydrate, or absent;

- R_{11} , R_{25} , can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO₂-), N₃-, NH₂-, MeSO₂NH-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- 5 - either R_{33} or R_{14} = alkyl group, and
the stereochemistry of R_{25} is in the β orientation;

k. Compounds of the above general formula III, wherein:

- $R_1 = R_2 = R_4 = R_5 = R_6 = R_7 = R_8 = R_{10} = R_{11} = R_9 = R_{12} = R_{13} = R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} =$
10 $R_{21} = R_{22} = R_{23} = R_{24} = R_{25} = R_{26} = R_{27} = R_{28} = R_{29} = R_{30} = R_{31} = R_{32} = R_{33} = H$,
- $R_{14} = CH_3$,
- $R_{34} = -OH$ or $-OR$ where R = alkyl, acyl or carbohydrate and
 $R_{35} = H$ or is absent
 $R_{37} = H$, $-OH$ or $=O$

- 15 $R_{36} = H$ or $-OH$

..... represents a single bond, and

- the methyl group at C25 may be either in the R or S configuration and

the stereochemistry of R_{25} is in the β orientation

wherein in addition to the above

- 20 at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, $=O$,
and OR where R = alkyl or acyl group or absent,
and X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO₂-), and

- 25 - N₃-, NH₂-, MeSO₂NH-

- alkyl;

l. Compounds of the above general formula III or carbohydrate derivatives thereof,
wherein:

- 30 - $R_1 = R_2 = R_4 = R_5 = R_6 = R_7 = R_8 = R_{10} = R_{11} = R_9 = R_{12} = R_{13} = R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} =$
 $R_{21} = R_{22} = R_{23} = R_{24} = R_{25} = R_{26} = R_{27} = R_{28} = R_{29} = R_{30} = R_{31} = R_{32} = R_{19} = R_{20} = H$,
- $R_{14} = R_{33} = CH_3$,

- $R_{34} = -OH$ or $-OR$ where R = alkyl, acyl or carbohydrate and

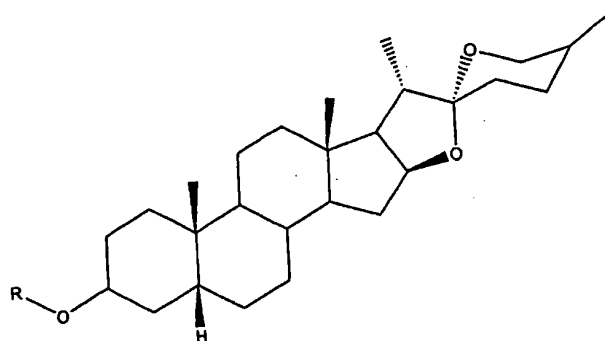
$R_{35} = H$ or is absent

- 35 $R_{37} = H$, $-OH$ or $=O$

$R_{36} = H$ or $-OH$

- represents a single bond, and
- the methyl group at C25 may be either in the R or S configuration and the stereochemistry of R₂₅ is in the β orientation wherein in addition to the above
- 5 at least one of R₃ or R₂₃ is a X radical, the possible remaining substituent being H, OH, =O, and OR where R = alkyl or acyl group or absent, and X is chosen from the group consisting of :
- halo atom,
 - (Me-S-), (Me-SO-), (Me-SO₂-), and
- 10 - N₃-, NH₂-, MeSO₂NH-
- alkyl;
- m. Substituted sapogenins wherein at least one OH-group of the sapogenin is substituted with X, chosen from the group consisting of :
- 15 - halo atom,
- (Me-S-), (Me-SO-), (Me-SO₂-),
 - N₃-, NH₂-, MeSO₂NH-, and
 - alkyl;
- 20 n. Sapogenins defined above wherein in the definition of X the halo atom is a fluoro atom;
- o. Substituted sapogenins selected from :
- 25 (3 β -fluoro-5 β , 20 α ,22 α ,25R-spirostane), (3,3-difluoro-5 β , 20 α ,22 α ,25R-spirostane), (3 α -methylsulphonylamino-5 β ,20 α ,22 α ,25R-spirostane), (3 α -azido-5 β , 20 α ,22 α ,25R-spirostane), (3 α -amino-5 β ,20 α ,22 α ,25R-spirostane), and their stereoisomers and racemic mixtures, their pharmaceutically acceptable pro-drugs and salts;
- p. Substituted sapogenins wherein the parent sapogenin which is then substituted with
- 30 at least one X radical as defined above is selected from sarsasapogenin, episarsasapogenin, smilagenin, epismilagenin, and anzurogenin-D;

- q. Compounds of the general formula Ia:



(Ia)

5

wherein the group R is selected from hydrogen; alkylcarbonyl; alkoxy carbonyl; alkyl-carbamoyl; or arylcarbonyl; or sulpho (HO_3S); phosphono ($(\text{HO})_2\text{P}(\text{O})-$); or a mono-, di- or tri-saccharide; wherein any alkyl group is optionally substituted with aryl, amino, mono- or di-alkyl-amino, a carboxylic acid residue ($-\text{COOH}$), or any combination thereof; and

10

- r. Derivative forms of the above compounds as defined as items a to q, in which the 3-position carbon atom or, in the case of Formulae II and III, the 3-position carbon atom, the 26-position carbon atom or each of the carbon atoms at the 3- and 26-positions, carries an O-sugar moiety wherein the sugar group is a mono-, di- or tri-saccharide, and acylated derivatives thereof.

15

3. A use according to claim 1 or claim 2, wherein the active agent, or at least one of the active agents, is selected from compounds of the general formula Ia.

20

4. A use according to any one of the preceding claims, wherein the active agent, or at least one of the active agents, is selected from:

sarsasapogenin

sarsasapogenin cathylate

sarsasapogenin acetate

25

sarsasapogenin succinate and pharmaceutically acceptable salts thereof

sarsasapogenin glycinate and pharmaceutically acceptable salts thereof

sarsasapogenin alaninate and pharmaceutically acceptable salts thereof

sarsasapogenin valinate and pharmaceutically acceptable salts thereof

- sarsasapogenin phenylalaninate and pharmaceutically acceptable salts thereof
sarsasapogenin isoleucinate and pharmaceutically acceptable salts thereof
sarsasapogenin methioninate and pharmaceutically acceptable salts thereof
episarsasapogenin
- 5 episarsasapogenin cathylate
episarsasapogenin acetate
episarsasapogenin succinate and pharmaceutically acceptable salts thereof
episarsasapogenin glycinate and pharmaceutically acceptable salts thereof
episarsasapogenin alaninate and pharmaceutically acceptable salts thereof
- 10 episarsasapogenin valinate and pharmaceutically acceptable salts thereof
episarsasapogenin phenylalaninate and pharmaceutically acceptable salts thereof
episarsasapogenin isoleucinate and pharmaceutically acceptable salts thereof
episarsasapogenin methioninate and pharmaceutically acceptable salts thereof
smilagenin
- 15 smilagenin cathylate
smilagenin acetate
smilagenin succinate and pharmaceutically acceptable salts thereof
smilagenin glycinate and pharmaceutically acceptable salts thereof
smilagenin alaninate and pharmaceutically acceptable salts thereof
- 20 smilagenin valinate and pharmaceutically acceptable salts thereof
smilagenin phenylalaninate and pharmaceutically acceptable salts thereof
smilagenin isoleucinate and pharmaceutically acceptable salts thereof
smilagenin methioninate and pharmaceutically acceptable salts thereof
epismilagenin
- 25 epismilagenin cathylate
epismilagenin acetate
epismilagenin succinate and pharmaceutically acceptable salts thereof
epismilagenin glycinate and pharmaceutically acceptable salts thereof
epismilagenin alaninate and pharmaceutically acceptable salts thereof
- 30 epismilagenin valinate and pharmaceutically acceptable salts thereof
epismilagenin phenylalaninate and pharmaceutically acceptable salts thereof
epismilagenin isoleucinate and pharmaceutically acceptable salts thereof
epismilagenin methioninate and pharmaceutically acceptable salts thereof.
- 35 saponin derivatives of sarsasapogenin, episarsasapogenin, smilagenin and epismilagenin in
which, in each case, the 3-position carbon atom carries an O-sugar moiety wherein the

sugar group is selected from glucose, mannose, fructose, galactose, maltose, cellobiose, sucrose, rhamnose, xylose, arabinose, fucose, quinovose, apiose, lactose, galactose-glucose, glucose-arabinose, fucose-glucose, rhamnose-glucose, glucose-glucose-glucose, glucose-rhamnose, mannose-glucose, glucose-(rhamnose)-glucose, glucose-(rhamnose)-rhamnose, glucose-(glucose)-glucose, galactose-(rhamnose)-galactose and acylated derivatives thereof;

16,22-epoxycoprostan-3 β -ol, smilagenone, coprosterol, and pharmaceutically acceptable pro-drugs and salts thereof.

10

5. A use according to any one of the preceding claims, wherein the active agent is present in a composition selected from pharmaceutical compositions, foodstuffs, food supplements and beverages.

15

6. A use according to any one of the preceding claims, wherein the active agent is present with one or more additional active agent.

7. A use according to claim 6, wherein the one or more additional active agent is selected from, but not limited, to cholinesterase inhibitors, dopamine agonists, COMT inhibitors, MAO-B inhibitors, anti-cholinergics, acetylcholine agonists, serotonin agonists, AMPA receptor agonists, GABA receptor agonists, NMDA receptor agonists, β -adrenoreceptor agonists, digoxin, dobutamine, anti-inflammatories, neurotrophic factors, statins, adenosine A2a receptor antagonists, aldose reductase inhibitors, immunomodulators, cannabinoid agonists, interferon β or tricyclic anti-depressants.

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8. A use according to any one of the preceding claims, wherein the human or non-human animal is suffering from, or is susceptible to, any of: Parkinson's disease, postencephalitic Parkinsonism, depression, schizophrenia, muscular dystrophy including facioscapulohumeral muscular dystrophy (FSH), Duchenne muscular dystrophy, Becker muscular dystrophy and Bruce's muscular dystrophy, Fuchs' dystrophy, myotonic dystrophy, corneal dystrophy, reflex sympathetic dystrophy syndrome (RSDSA), neurovascular dystrophy, myasthenia gravis, Lambert Eaton disease, Huntington's disease, motor neurone diseases including amyotrophic lateral sclerosis (ALS), multiple sclerosis, postural hypotension, traumatic neurodegeneration e.g. following stroke or following an accident (for example, traumatic head injury or spinal cord injury), Batten's disease, Cockayne syndrome, Down syndrome, corticobasal ganglionic degeneration, multiple

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system atrophy, cerebral atrophy, olivopontocerebellar atrophy, dentatorubral atrophy, pallidoluysian atrophy, spinobulbar atrophy, optic neuritis, sclerosing pan-encephalitis (SSPE), attention deficit disorder, post-viral encephalitis, post-poliomyelitis syndrome, Fahr's syndrome, Joubert syndrome, Guillain-Barre syndrome, lissencephaly, Moyamoya disease, neuronal migration disorders, autistic syndrome, polyglutamine disease, Niemann-Pick disease, progressive multifocal leukoencephalopathy, pseudotumor cerebri, Refsum disease, Zellweger syndrome, supranuclear palsy, Friedreich's ataxia, spinocerebellar ataxia type 2, Rhetts syndrome, Shy-Drager syndrome, tuberous sclerosis, Pick's disease, chronic fatigue syndrome, neuropathies including hereditary neuropathy, diabetic neuropathy and mitotic neuropathy, prion-based neurodegeneration, including Creutzfeldt-Jakob disease (CJD), variant CJD, new variant CJD, bovine spongiform encephalopathy (BSE), GSS, FFI, kuru and Alper's syndrome, Joseph's disease, acute disseminated encephalomyelitis, arachnoiditis, vascular lesions of the central nervous system, loss of extremity neuronal function, Charcot-Marie-Tooth disease, susceptibility to heart failure, asthma, and macular degeneration.